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Historical Vignette

Management of basilar invagination: A historical perspective

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Abstract

For a long time the terms basilar invagination and platybasia were used interchangeably. Basilar invagination has been defined as a prolapse of the vertebral column into the spinal cord. Platybasia is defined as an abnormal obtuse angle between the anterior skull base and the clivus. The authors review the existing literature and summarize the historical and modern perspectives in the management of basilar invagination. From radiological curiosities, the subject of basilar invagination is now viewed as eminently treatable. The more pronounced understanding of the subject has taken place in the last three decades when on the basis of understanding of the biomechanical subtleties the treatment paradigm has remarkably altered. From surgery that involved decompression of the region, stabilization and realignment now form the basis of treatment.

Key words: Basilar invagination, facetal distraction, Goel's craniovertebral realignment, history, intra-articular spacers

BASILAR INVAGINATION: FROM RADIOLOGICAL AND PATHOLOGICAL CURIOSITIES TO A TREATABLE ENTITY — A HISTORICAL VIEW POINT

Ackermann in 1790 described basilar invagination and a small posterior fossa in cretins.[1] Since this deformity at the base of the skull was more commonly seen in cretins, it was initially thought to be the cause of cretinism. According to Spillane, one of the first descriptions of basilar invagination was given by Anders Adolph Retzius and Frederik Theodor Berg in 1855.[2] They called it "*Impressio baseos crania ex atrophia adiposa (cerea).*" When the skull is viewed looking upwards from the caudal direction this abnormality appears as an impression of the base of the skull giving it its name.

The term "Platybasia" an abnormal flattening of the base of the skull was first coined by Virchow in 1857.[3] However, in 1876, Virchow after studying the anthropology of Frisians showed that basilar invagination was sometimes seen with platybasia.[4] In association with a flattening of the base of the skull there was an upward displacement of the basilar and condylar portions of the occipital bone that caused infolding of the foramen magnum, reduction of the posterior fossa, and consequent protrusion of

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the upper cervical spine into the cervicomedullary junction with resultant neurological signs.

Most anatomists at the time thought that this deformation had a mechanical cause hence the name was applied to it, but Virchow and more particularly Grawitz suggested that the condition was largely the result of a developmental anomaly of the bone.[5,6] From the eighteenth century to the twenty-first century we have come full circle with current thinking attributing the cause of basilar invagination to be mechanical as we will see later.

Chamberlain discussed basilar impression (platybasia) as a deformity of the occipital bone and upper end of the cervical spine resulting from anomalous development.[7] In his paper, he describes basilar impression as "it is as though the weight of the head has caused the ears to approach the shoulders, while the cervical spine, refusing to be shortened, has pushed the floor of the posterior fossa upward into the brain space."

At this time anatomic studies were convincing to the effect that basilar impression is a developmental anomaly as there were no indications of any other disease process, such as rickets or osteomalacia, visualized. This developmental theory however failed to explain the appearance of neurological symptoms and signs in the later decades of life. The explanation offered for this was that the young and developing brain was more tolerant to the compressive effects that later proved deleterious to older tissues. However, Dr. Richard Light was of the opinion that the increase in length of the spine during the normal growth spurt may produce traction on the cord that has already attained its normal length and thus cause pressure of the odontoid process on the medulla and produce symptoms.[7]

For a long time basilar invagination remained an anatomic and radiological curiosity and it is only in recent years that this entity has entered the realm of clinical neurosurgery.

It was suggested by many that as basilar invagination has several etiologies it should be considered a radiographic finding and the underlying etiology should be identified. Among the reported etiologies of basilar invagination are clival hypoplasia, condylar hypoplasia, hypoplasia of the atlas, incomplete ring of atlas with spreading of the lateral masses, achondroplasia, and atlanto-occipital assimilation. "Basilar impression" is the term used to describe the acquired form of basilar invagination, which results from softening of the bone at the base of the skull. Common conditions leading to basilar impression are Paget's disease, osteomalacia, hyperparathyroidism, osteogenesis imperfecta, Hurler syndrome, rickets, and skull base infection.

Most of the early reports of basilar invagination were based on postmortem findings, till Schuller in 1911 made the first radiological diagnosis in a living patient.[8] The radiological diagnosis was later refined by Chamberlain and other radiologists and various parameters were defined to make the diagnosis of basilar invagination. The lines used by them for diagnosis were as follows:

1. Chamberlain's Line — The tip of the dens is >5 mm above a line joining the posterior edge of the hard palate to the dorsal lip of the foramen magnum.
2. McGregor's Line — The tip of the dens is >7 mm above the back of the hard palate to the lowest point of the occipital squama.
3. Bull's angle — The intersection of the hard palate and the plane of the atlas; 13 degrees is accepted as the upper limit of normal.
4. Fischgold's digastric line—Line joining the digastric grooves of the mastoid processes on anteroposterior X-ray or transoral tomogram; normally passes through the atlanto-occipital articulation.

BASILAR INVAGINATION — EARLY TREATMENT

In 1939, Chamberlain reported four patients with basilar invagination. He recommended suboccipital craniectomy with cervical laminectomy and dural opening for the management of these patients.[7] The treatment was based on relieving the compression on the cervicomedullary junction. Even in those early times, A. De Vet questioned the utility of suboccipital decompression.[9] He was of the opinion that the decompression may only be symptomatic treatment and one must consider the possibility of acceleration of the disease after removal of a large portion of the occipital bone. Under normal conditions, the bone that is removed has no supporting function but in basilar impression the normal planes of support fail and maybe replaced by the posterior arch of atlas and the foramen magnum. Sekir suggested the use of traction along with a supporting apparatus in such cases till the progress of the disorder has halted. The suboccipital decompression could be reserved only for those patients who show signs of raised intracranial pressure.[9] In the subsequent years, more reports of patients with craniovertebral anomalies followed and the treatment consisted of posterior fossa decompression by enlargement of the foramen magnum and removal of the posterior arch of the C1 vertebra. The morbidity and mortality in these patients however remained high. In 1964, Barucha and Dastur in their paper on craniovertebral anomalies brought out the significant morbidity and mortality in the patients who underwent a dorsal decompressive procedure for these abnormalities.[10] Dastur and Sinh reported a series of patients who developed intramedullary hemorrhage following posterior decompression surgery and questioned the validity of this form of treatment.[11]

In 1980, Menezes *et al.* proposed a treatment algorithm for craniocervical junction abnormalities.[12,13] They divided craniocervical abnormalities into reducible and irreducible groups. For reducible pathologies they recommended posterior fixation. Irreducible pathologies were further divided on the basis of site of compression into ventral and dorsal groups. For ventral stable pathologies only a transoral decompression was carried out and for ventral unstable pathologies a transoral decompression was followed by posterior occipitocervical

fixation. Similarly for dorsal pathologies a dorsal decompression with or without stabilization was carried out. The algorithm of the proposed treatment format as proposed by Menezes *et al*. continues to be popular. The atlantoaxial and occipitoaxial fixation techniques developed and evolved. From only bone overlay to sublaminar wire fixation and subsequently screw plate/rod fixation techniques have become popular.[14] From midline fixation methods the focus has now shifted to facetal fixation.[15-17] Currently, the technique proposed by Goel and Laheri in the year 1988 and published in 1994 is the most accepted technique of atlantoaxial stabilization.[15]

Over the years there has been an increased understanding of the clinical presenting profile of these patients. More importantly, introduction of computer-based imaging clarified the anatomical subtleties of the region. Anatomic and biomechanical studies helped further in evaluating and treating these complex and surgically challenging cases.

Goel *et al*. in 1998 divided basilar invagination into two groups on the basis of presence or absence of Chiari malformation.[18] The prime issue in this classification was the understanding that atlantoaxial dislocation in both groups was considered to be of fixed or of irreducible variety. Essentially, Group I included cases where there was invagination of the odontoid process into the foramen magnum and it indented into the brainstem. The tip of the odontoid process distanced itself from the anterior arch of the atlas or the inferior aspect of the clivus. The angle of the clivus and the posterior cranial fossa volume were essentially unaffected in these cases. In Group II cases, on the other hand, the assembly of odontoid process, anterior arch of the atlas and the clivus migrated superiorly in unison resulting in reduction of the posterior cranial fossa volume, which was the primary pathology in these cases. The Chiari malformation or herniation of the cerebellar tonsil was considered to be a result of reduction in the posterior cranial fossa volume.[18] In Group I, basilar invagination the tip of the odontoid process "invaginated" into the foramen magnum and was above the Chamberlain line,[7] McRae line of foramen magnum[19] and Wackenheim's clival line.[20] The definition of basilar invagination of prolapse of the cervical spine into the base of the skull, as suggested by von Torklus,[21] was suitable for this group of patients. Group II basilar invagination was where the odontoid process and clivus remained anatomically aligned despite the presence of basilar invagination and other associated anomalies. In this group, the tip of the odontoid process was above the Chamberlain's line but below the McRae's and the Wackenheim's lines. In the year 1997, Goel first defined the clinical implication of association of small posterior cranial fossa volume and Chiari malformation. As deformity rather than instability was considered to be the issue in pathogenesis, decompression of the neural structures rather than stabilization was considered to be the therapeutic goal. On the basis of this study, Goel recommended transoral decompression with or without atlantoaxial or occipitocervical fixation in Group I patients and foramen magnum decompression in Group II patients. Goel also suggested that opening of the dura during posterior fossa decompression was not recommendable and should be avoided in all cases, including those where there was an association of Chiari malformation and syringomyelia.[18] Prior to this dural opening and widening of posterior fossa by dural graft placement was an accepted norm.

However, not all questions were answered based on this strategy and the entity of basilar invagination was still an enigma.

BASILAR INVAGINATION — REVOLUTIONARY CHANGE IN MANAGEMENT

All of the previous treatment philosophies were based on the premise that basilar invagination is a developmental and a "fixed" or a stable anomaly and neural compression is the cause of symptoms.

On further evaluation and understanding of the abnormality Dr. Goel reported his experience of treatment of specific subgroup of basilar invagination by atlantoaxial joint distraction using bone graft and intra-articular spacers and direct lateral mass fixation.[22] This paper initiated a change in the understanding of the subject and caused a paradigm shift in the management of basilar invagination. The concept that changed is that the atlantoaxial joint in a select group of cases is not only not fixed but is mobile and in many cases hypermobile. More importantly, the concept that the atlantoaxial dislocation can be reduced in these cases and the craniovertebral junction can be realigned revolutionized the treatment strategies in these cases.

In this paper, basilar invagination was divided into two groups.[22] In one subgroup of patients there was clear radiological evidence of instability of the region that was seen as distancing of the odontoid process away from the anterior arch of atlas/clivus or the atlantodental or clivodental interval was abnormally increased. The tip of the odontoid process was above the Chamberlain line, McRae line and Wackenheim clival line. This subgroup of patients was labeled as having Group A basilar invagination. The radiological findings suggested that the odontoid process in Group A patients resulted in direct compression of the brainstem. In some Group A patients, there was Chiari 1 malformation, and this feature differentiated the present classification from the earlier classification. The pathogenesis in patients with Group A basilar invagination was mechanical instability of the region. In these patients, the atlantoaxial joints were in an abnormally inclined or oblique position instead of the normally found horizontal orientation. The alignment of facets of atlas and facets of axis simulated positioning of vertebral bodies in lumbosacral listhesis.[23] Basilar invagination in this subgroup of patients appeared to be related to progressively increasing listhesis of facets of atlas over the facets of axis. The joint in these cases is not fixed or fused but is mobile or hypermobile, thus is probably the primary cause of basilar invagination. As instability or listhesis is the cause of basilar invagination, stabilization as recommended for lumbosacral listhesis formed the baseline of surgical treatment.[23] This theory also explains the appearance

of neurological symptoms and signs in these patients at a later age group and also the increase in symptoms following trauma. The basilar invagination could be reduced *by "playing with the atlantoaxial joint"* The technique involved *manipulation, distraction, reduction, and fixation* of the atlantoaxial joint. The distraction and reduction was maintained with the help of bone graft and/or spacers. The suggestion of use of intra-articular spacers for the purpose of distraction of the facets, reduction of the basilar invagination, stabilization of the joint, and its realignment altered the treatment concept of basilar invagination. The fixation was performed with Goel's lateral mass plate and screw fixation technique.[15,16] In Group B, the entire complex of the clivus, basiocciput and the craniovertebral junction was rostrally located and the tip of the odontoid process was superior to the Chamberlain line but inferior to the McRae and the Wackenheim lines. In Group B, the pathogenesis appeared to be congenital dysgenesis of the region. The atlantoaxial joint was considered to be stable or fixed and instability was not considered an issue in this group of patients. Foramen magnum decompression was identified to be the treatment for Group B basilar invagination, as small posterior cranial fossa volume was identified to be the pathological issue in this group.

Thus, began the era of "craniovertebral realignment" for basilar invagination. The key points in treatment were reduction of the basilar invagination and atlanto-axial fixation. The stabilization of the region was more important than the degree of reduction.

For a long time, however, the combined surgery of transoral decompression followed by occipito-cervical fixation was continued and it is only in recent times that neurosurgeons have begun to understand the philosophy and are attempting 'posterior only' method of reduction and fixation for basilar invagination. Recently, there have been some modification of the distraction reduction procedures suggested that includes the occiput in fixation.[24,25] However, opening of the atlantoaxial joint and direct fixation forms the current Gold standard of treatment. Inclusion of the occiput may not only be unnecessary but maybe a suboptimal construct biomechanically. Goel's technique of distraction and fixation of the C1–C2 joints can lead to reduction of basilar invagination and realignment of the craniovertebral junction.

BASILAR INVAGINATION — RECENT THINKING

In the year 2014, as the understanding of the entity of basilar invagination has evolved, Goel *et al*. have identified that atlantoaxial instability is the nodal point of pathogenesis of all types of basilar invagination including Group II and Group B basilar invagination. This instability may be identified on the basis of facetal malalignment on radiological imaging.[26-28] Three types of facetal malalignment can be observed; Types I, II, and III. In Type 1 facetal instability, wherein the facet of atlas is displaced anterior to the facet of axis, (more frequently identified with Group 1 and Group A basilar invagination), the basilar invagination is usually identified in younger patients and is associated with more acute clinical symptoms. The odontoid process is displaced posteriorly and compresses the neural structures. On the other hand, basilar invagination related to Type 2 (facet of atlas is displaced posterior to the facet of axis) and Type 3 facetal instability, wherein the facets of atlas and axis are in alignment and instability can be identified only by direct facetal manipulation during surgery, is more frequently identified with Group II and Group B basilar invagination. Type 3 facetal instability is also called central or axial atlantoaxial instability. Types 2 and 3 instability related basilar invagination is identified in relatively older patients and is associated with more chronic or longstanding structural malformations. It is now believed that atlantoaxial fixation forms the basis of treatment in all types of basilar invagination. Decompression of bone either by transoral route or by posterior foramen magnum decompression are suboptimal forms of treatment.

BASILAR INVAGINATION AND ASSOCIATED ABNORMALITIES

A number of bone and soft tissue anomalies are associated with basilar invagination. These include short neck, torticollis, platybasia, cervical vertebral body fusion (Klippel-Feil abnormality) including assimilation of atlas, spondylotic spinal changes, and restriction of neck movements. A number of these abnormalities were seen to be reversible following decompression and stabilization of the region.[29] Considering that several physical features associated with this group of basilar invagination are reversible, it appears that the pathogenesis in such cases may be more due to mechanical and secondary factors rather than congenital causes or embryological dysgenesis. The common teaching on the subject is that the short neck and torticollis are a result of embryological dysgenesis and effectively result in indentation of the odontoid process into the cervicomedullary cord. However, it appears that atlantoaxial dislocation and its related cord compression due to indentation by the odontoid process is the primary event and all the physical alterations and bony abnormalities, including the short neck and torticollis, are secondary natural protective responses that aim to reduce the stretch of the cord over the indenting odontoid process. Pain, restriction of neck movements, and hyperlordosis of the neck indicate the presence of instability of the craniovertebral junction. All of these natural responses probably allow the cord a relatively stretch-free traversal over the indenting odontoid process. Reduction of the disc spaces, osteophytes formation, incomplete and complete cervical fusions, and alterations in the craniospinal and cervical angulations appear to be directly related to the reduction in neck length. The reduction in the disc-space height and fusions are more prominently seen in the upper cervical vertebrae. It appears that cervical fusions and assimilation of the atlas may be related to long-standing and progressive reduction in the disc-space height. The structural alterations are external manifestations of naturally protective responses that aim to stall the effects and consequences of instability. Chiari 1 malformation and syringomyelia are internal or neural manifestations also designed

as protective responses related to atlantoaxial instability and are aimed at delaying or stalling the neurological sequelae.[30,31]

ROLE OF TRANSORAL SURGERY

Transoral surgery first began when Kanavel in 1917 used this approach to remove a bullet from the craniocervical junction.[32] It gained popularity in the 1980s when Crockard *et al.* started using the approach to access the craniovertebral junction with the aid of retractor systems and the microscope.[33] The approach continued to remain the workhorse of craniovertebral junction surgery till recently. Following the concept of craniovertebral realignment as pioneered by Goel *et al.*, the indications of transoral surgery in cases of basilar invagination are becoming fewer. The current surgical concept is based on the understanding that basilar invagination is a result of atlantoaxial instability and atlantoaxial stabilization forms the bottom-line of treatment. Any form of decompression, either transoral or foramen magnum decompression, is not necessary and may be countereffective in the long run.

CONCLUSIONS

As the understanding of basilar invagination has evolved it appears that mechanical instability is the cause and craniovertebral realignment and atlantoaxial fixation is the current management of this anomaly. The key is exposure, manipulation, and stabilization of the atlantoaxial joints. Inclusion of the occiput and subaxial spine in the fixation construct is not necessary and maybe suboptimal.

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